# CASE REPORTS

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## Bilateral Eventration of the Diaphragm

MICHAEL H. WELLER, MD
*Long Beach*

DIAPHRAGMATIC EVENTRATION REFERS to absence or deficiency of diaphragmatic muscle, with resultant elevation. Bilateral diaphragmatic eventration is uncommon, and the prognosis is usually poor. The purpose of this report is to describe three cases and to review the roentgenographic appearance of bilateral eventration and the literature.

### Reports of Cases

Case 1. A 2,180-gram boy was born at 37 weeks gestation after an otherwise uncomplicated pregnancy. There was immediate respiratory distress. The Apgar sore was 1 and 4 at one and five minutes respectively. Following endotracheal intubation, high ventilatory pressures were required. No bowel sounds were heard in the chest. The abdomen was scaphoid. Diaphragmatic hernia was suspected.

Results of initial arterial blood gas analysis included oxygen pressure ($Po_2$) of 49, an arterial carbon dioxide pressure ($Pco_2$) of 52 and a pH of 7.10. Arterial oxygen saturation was 75 percent. On a roentgenogram of the chest, pronounced elevation of both hemidiaphragms was seen, consistent with bilateral eventration. The stomach was in normal position (Figure 1). Despite maximum respirator settings, an arterial $Po_2$ above 50 could not be obtained. Right diaphragmatic plication was carried out at several hours of age. Postoperatively, the right lung would not expand to fill the pleural space (Figure 2). The infant died at 19 hours of age. Findings on postmortem examination confirmed elevation of both hemidiaphragms. Histologic examination of the diaphragm showed only collagen, with no muscle fibers. The lungs were hypoplastic and weighed a total of 10 grams. Microscopically, bronchi and alveolar ducts were seen, with only rare alveoli. Hyaline membranes lined many alveolar ducts. The vessels were thick and immature. Focal hemorrhage was present.

CASE 2. A 2,700-gram boy was born after a pregnancy complicated by mild polyhydramnios and premature rupture of membranes. The Apgar score was 5, both at birth and at three minutes. There was immediate grunting and cyanosis. Chest wall motion and lung ventilation were diminished, and there was obvious use of accessory respiratory muscles. Findings on arterial blood gas analysis showed a $Po_2$ of 55, a $Pco_2$ of 74 and a pH of 7.16. On radiographs of the chest, persistent right-sided diaphragmatic elevation was noted (Figure 3). Fluoroscopic examination showed restricted motion of both hemidiaphragms, but no para-

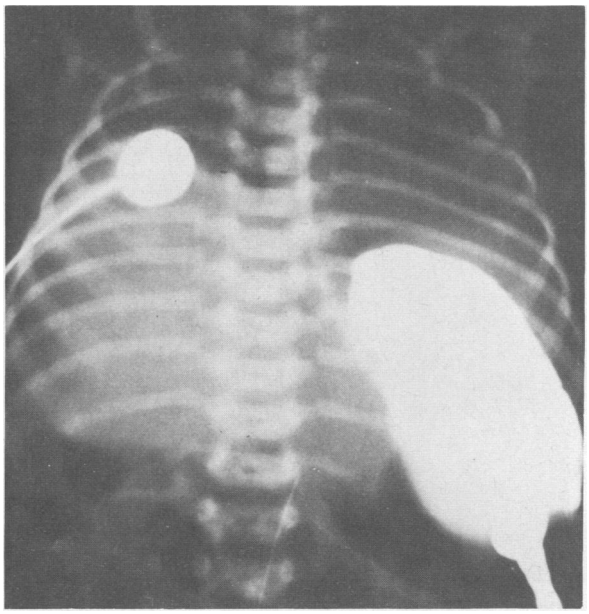

**Figure 1.—(Case 1) Bilateral elevation of the diaphragm is present. The elevated dome of the left hemidiaphragm can be seen faintly just above the barium-filled fundus.**

From the Department of Radiology, University Hospital, University of California, San Diego, School of Medicine, San Diego.

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Reprint requests to: Michael Weller, MD, Department of Radiology, Memorial Hospital Medical Center of Long Beach, P.O. Box 1428, Long Beach, CA 90801.

doxical motion. Progressive atelectasis of the left lower lobe occurred despite manual ventilation. On day 14, a right diaphragmatic plication was carried out with only mild initial improvement, and subsequent diaphragmatic elevation. The infant died on day 30. Results of postmortem examination showed bilateral diaphragmatic eventration, bilateral pneumonia and interstitial fibrosis. There was no pulmonary hypoplasia. Findings were within normal limits on histologic examination of the brain stem, spinal cord, peripheral nerve and skeletal muscle.

CASE 3. A 2,948-gram boy was born by cesarean section for nonprogressive labor and ruptured membranes. Apgar scores were 3 and 5 at one and five minutes. Cyanosis, grunting, decreased thoracic excursion and decreased basilar breath sounds were noted. On arterial blood gas analysis, there was a $Po_2$ of 59, a $Pco_2$ reported at greater than 100 and a pH of 7.02. On a radiograph of the chest, elevation of both hemidiaphragms and bilateral lower lobe atelectasis were seen (Figure 4). The ribs and clavicles were thin, but there were no rib fractures. Skeletal survey disclosed overconstriction of the shafts of the tubular bones and diminished muscle bulk. Progressive central nervous system dysfunction occurred. A diagnosis of arthrogryposis multiplex congenita was made. Findings from a skeletal muscle biopsy were "compatible with neurogenic atrophy." The infant died at 15 days of age. On postmortem examination, bilateral eventration of the diaphragm was noted, worse on the right, which consisted mostly of fibrous tissue. The lungs weighed slightly less than normal and showed a mild lack of development of the periphery. Vessels and alveolar ducts extended to within two or three avleoli of the pleura. Skeletal muscle showed pronounced fibrous replacement. The brain stem, spinal cord and peripheral nerve were found to be normal.

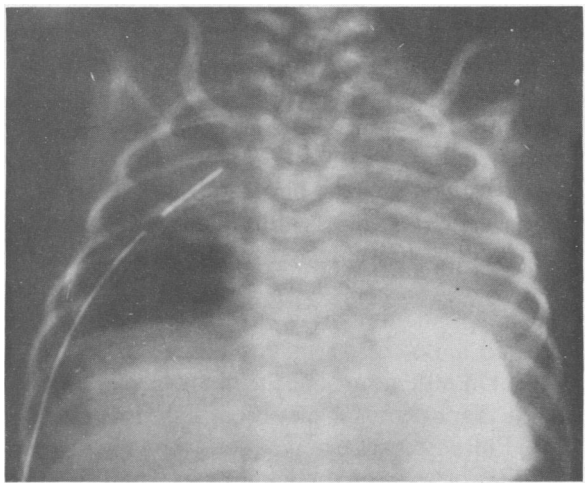

**Figure 2.**—(Case 1) Postoperative radiograph of the chest made following right diaphragmatic plication. There is failure of expansion of the hypoplastic right lung.

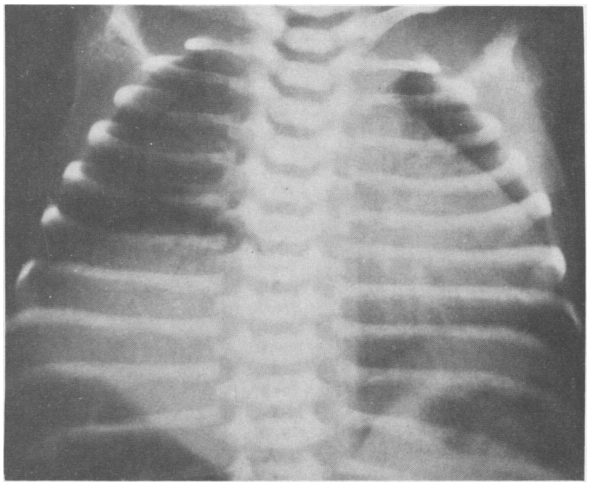

**Figure 3.**—(Case 2) Radiograph of the chest made during the first 24 hours of life showing elevation of the right hemidiaphragm. The left hemidiaphragm is not elevated but at autopsy was found to contain no muscle.

## Discussion

Eventration refers to diaphragmatic elevation due to muscular deficiency. It is more often unilateral, and may present at any age.* Unilateral

*References 3-5,8,9,11-13,15,19-21,25-30,32,34,35

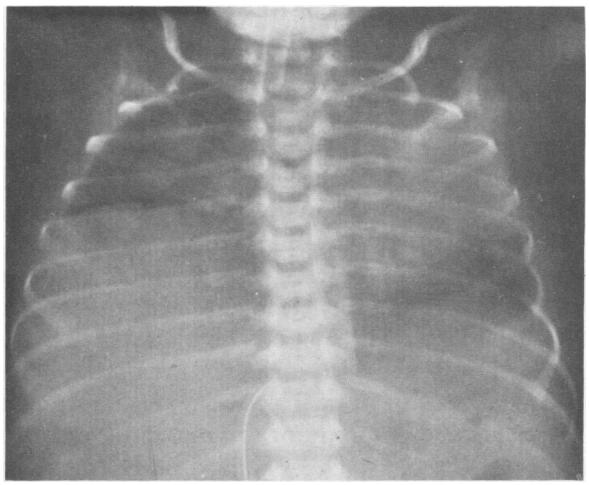

**Figure 4**—(Case 3) Radiograph of the chest made at 12 hours of age shows right diaphragmatic elevation. On subsequent radiographs, left sided elevation was seen as well. There is atelectasis involving both lungs. The ribs and clavicles have a slender, poorly ossified appearance.

TABLE 1.—*Review of Cases of Bilateral Eventration*

| *Author(s)* | *Age* | *Sex* | *Onset of Symptoms* | *Symptoms* | *Radiologic Findings* | *Surgical Procedures* | *Pulmonary Hypoplasia* | *Comment* |
|---|---|---|---|---|---|---|---|---|
| Landon[19] 1936 | 4 wks | Male | Birth | Cyanosis, dyspnea | Chest x-ray: elevation of right hemidiaphragm. Fluoroscope: diminished diaphragmatic excursion on the right; normal excursion on the left. | No | .... | Died day 80. Autopsy: right hemidiaphragm-peripheral muscle only. Left hemidiaphragm central fibrous membrane without muscle. Anomalous right lung fissures. |
| Avnet[1] 1962 | 5 days | Male | Birth | Transient respiratory distress, cyanosis (but had meconium staining) | Bilateral anterior eventration (pneumoperitoneography) | No | .... | Died age 6 weeks. Autopsy: Potter's facies. Multiple congenital anomalies. Cerebral toxoplasmosis. Bilateral anterior eventration. |
| | 10 mos | Male | .... | Asymptomatic | Bilateral anterior eventration (pneumoperitoneography) | No | .... | No follow-up. Angiocardiogram-coarctation aorta (mild) |
| Lundstrom and Allen[22] 1966 | 5 wks | ? | .... | Asymptomatic | Chest x-ray: elevation of left hemidiaphragm. Pneumoperitoneography: bilateral eventration | Yes; bilateral (staged) | .... | Surgical findings: Diminished muscle anteriorly and laterally. ? absent spleen. |
| Firestone and Taybi[14] 1967 | 5 mos | ? | 5 mos. | Shortness of breath. Feeding difficulties | Bilateral anterior eventration (pneumoperitoneography) | Yes; bilateral | .... | Died postoperatively. Autopsy: Bilateral anterior eventration, chronic pulmonary inflammation. |
| Wayne et al[37] 1973 | 3 days | Female | Birth | Labored respirations, mild cyanosis | X-ray film of chest: elevation of both hemidiaphragms. Bilateral pneumonia. Fluoroscope: diminished excursions of both hemidiaphragms | No | .... | Died day 12. Autopsy: Congenital cytomegalovirus infection-urine and cerebrospinal fluid. Almost total aplasia of skeletal muscle of diaphragm with muscle fibers at periphery only. "Normal" phrenic nerves. |
| Briggs et al[7] 1973 | 2 hrs | Female | Birth | Respiratory distress, cyanosis | Elevation of right hemidiaphragm. "Small lungs," basal atelectasis | No | .... | Died day 19. Autopsy: diaphragm shows no sign of muscle. Bilateral pulmonary hypoplasia. Congenital rubella. |
| Mellins et al[24] 1974 | 5 wks | Male | 4 wks | Cyanosis, tachypnea | Bilateral anterior eventration | Yes | .... | Died as 4 mos. Autopsy: eventration bilateral. Werdnig-Hoffmann disease. |
| | 2 mos | Male | 7 wks | Respiratory distress, cyanosis | Bilateral anterior eventration | Yes | .... | Died at 5 mos. Autopsy: eventration bilateral. Werdnig-Hoffmann disease. |
| Weller 1975 | Newborn | Male | Birth | Respiratory distress | Bilateral complete eventration | Yes | Yes | Died at 19 hours. Autopsy: No diaphragmatic muscle. Pronounced bilateral pulmonary hypoplasia. |
| | Newborn | Male | Birth | Respiratory distress, cyanosis | X-ray film of chest: elevation of both hemidiaphragms, right greater than left. Fluoroscope: diminished excursion bilaterally | Yes | .... | Died at 1 mo. Autopsy: diaphragm contains no muscle on right, little muscle on left. Spinal cord, peripheral nerve normal. |
| | Newborn | Male | Birth | Respiratory distress, cyanosis | Bilateral complete eventration, basal atelectasis. Diminished peripheral muscle mass; thin gracile ribs and long bones | No | Mild | Died at 13 days. Clinical diagnosis: arthrogryposis multiplex congenita. Autopsy: absence of diaphragmatic muscle on right. Diminished muscle on left. Normal spinal cord, peripheral nerves. Atrophic muscles. |

eventration is more common in males, and on the left side.[11,19,20,27,30,32] similar to diaphragmatic hernia. Respiratory distress is the usual presentation in the neonate.

Eventration is often confused with diaphragmatic hernia clinically and roentgenographically. The two may be impossible to differentiate surgically,[6,11] especially when abdominal contents are contained in a "sac" of parietal pleura and parietal peritoneum.[18] When eventration is present, there is an intermediate fibromuscular layer representing rudimentary diaphragm which may be difficult to identify. The clinical course is variable, and depends on the amount of diaphragmatic muscle and promptness of diaphragmatic plication. The prognosis in infants with unilateral eventration is guarded,[3,19-21,25,26,35] comparable to that of babies with acquired phrenic nerve paralysis.[11,15-17,29,33]

The details of the nine previous cases of bilateral eventration appear in Table 1. The three cases reported are similar in clinical and roentgenologic features. Respiratory distress and cyanosis began at birth. Diaphragmatic elevation was seen on radiographs of the chest. In two of the three infants, the right hemidiaphragm was more elevated than the left. Basal atelectasis was seen in two babies. Fluoroscopically, in the patient in Case 2 decreased diaphragmatic excursion bilaterally was seen. All three patients died, despite unilateral diaphragmatic plication in Cases 1 and 2. At postmortem examination, the diaphragm in Case 1 was found to have no muscle. The diaphragm in Cases 2 and 3 showed no muscle on the right and diminished muscle on the left. Of additional interest in Case 3 is the clinical diagnosis of arthrogryposis multiplex congenita, and the muscle biopsy consistent with neurogenic atrophy.

The cause of death in this disorder is unclear. Pulmonary hypoplasia is not a consistent postmortem finding. (Table 1). The lungs in Case 1 were severely hypoplastic. This baby had total absence of diaphragmatic muscle. There was no pulmonary hypoplasia in Case 2, and only mild hypoplasia in Case 3. With *partial* eventration, the cause of death is probably pneumonia, rather than pulmonary hypoplasia.

## Etiology

The cause of eventration is unknown. A defect in embryogenesis is suggested by the presence of eventration in newborns and the fetus[2,23] and occurrence of frequent coincident anomalies.* Associated ipsilateral cephalad ectopia of the kidney,[8] and other abdominal organs,[5,36] similar to diaphragmatic hernia, support a defect in embryogenesis. The mechanism of muscularization of the primitive diaphragm is not clear. Diaphragmatic muscle may arise *in situ* from condensed mesenchyme.[38] Muscularization may also occur from posteriorly by invasion of cervical myotomes migrating with intact nerve supply.[19,26,35] If this process fails, absence of diaphragmatic muscle or incomplete anterior muscularization could result. Mechanical interference with bronchiolar differentiation could then eventuate in pulmonary hypoplasia. Finally, as the pleural spaces enlarge, chest wall muscle may contribute to muscular investment of the diaphragm.[6] Incomplete development of pleural spaces might lead to peripheral muscularization only, but cannot explain anterior eventration. Failure of myotome migration may lead to anterior eventration. Most patients with anterior muscle deficiency have been relatively asymptomatic, and have a longer survival.[1,14,22,24,27,35] In babies with early respiratory distress, there either is no diaphragmatic muscle or only peripheral muscle.[13,19,20,25] If failure of pleural "burrowing" is the responsible mechanism in these patients, pulmonary hypoplasia could be primary, rather than secondary.

Failure of peripheral nerve development may result in eventration.[5] The phrenic nerve has been noted to be absent,[15] small or normal in various reports.[5,11,25,30,32] Partial eventration occurs with arthrogryposis multiplex congenita,[12] and a disease in sheep, similar to human arthrogryposis, shows evidence of muscular aplasia of the diaphragm.[2] Two patients with bilateral anterior eventration subsequently died of central nervous system disease at five and seven months of age.[24] In Cases 2 and 3, the brain, cervical cord and peripheral nerves showed no evidence of anterior-horn cell disease. In Case 3, findings in a premortem skeletal muscle biopsy were "compatible with neurogenic atrophy."

Congenital infection has been linked with eventration. There are reports of associated congenital rubella,[7] cytomegalovirus infection[37] and cerebral toxoplasmosis.[1] Perhaps viral or parasitic infection in utero affects central or peripheral nervous development resulting in neurogenic atrophy of the diaphragm. Death may occur before development of peripheral muscle disease.

*References 1,4,11,20,21,25,32,35

## Summary

The three cases reported are similar radiographically and confirm the uniformly dismal prognosis of bilateral eventration when symptoms are present at birth. The cause is probably a defect in embryogenesis, although the roles of central and peripheral diaphragmatic innervation, and of congenital infection are still unclear.

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# Mitral Valve Vegetation Simulating Left Atrial Myxoma

DAVID S. HIRSCHFELD, MD
BIANKA B. EMILSON, Tech Eng (Sweden)
*San Francisco*

THE ECHOCARDIOGRAPHIC appearance of vegetations in patients with endocarditis was first described by Dillon and associates in 1973.[1] Several subsequent reports have substantiated the important role of echocardiography in the diagnosis of endocarditis involving the aortic valve.[2-4] Considerably less has been written on the echocardiographic pattern of mitral valve vegetations. We recently saw a patient with mitral valve endocarditis in whom findings on an echocardiogram initially suggested the presence of a left atrial myxoma. The subsequent disappearance of the abnormal echoes concomitant with effective medical treatment of the endocarditis suggests that the mass of echoes noted represented a vegetation attached to the mitral valve.

From the Medical Service, San Francisco General Hospital, and the Department of Medicine, University of California, San Francisco.

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Reprint requests to: David S. Hirschfeld, MD, c/o Editorial Office, San Francisco General Hospital, 1001 Potrero Avenue, San Francisco, CA 94110.